

Anterior clinoid mucocoele presenting as progressive fluctuating visual loss mimicking inflammatory optic neuropathy

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Abstract:

Optic nerve lesions usually produce monocular visual loss, and in compressive optic neuropathies, gradual vision loss is the more usual presentation. Mucocele in the anterior clinoid process is a rare cause of compressive optic neuropathy that can lead to monocular visual loss. A 19-year-old Sri Lankan girl presented with progressive, painless fluctuating right-sided monocular visual loss over 1-year duration. On presentation, right side visual acuity was 6/60 and fundoscopy revealed pale disk on the right side. Ocular tomography showed right-sided temporal retinal thinning. Magnetic resonance imaging revealed right anterior clinoid process mucocele causing compression and kinking of the right optic nerve. Because the right side, optic disc was already pale and retina was thinned, we decided to manage conservatively. Early imaging in compressive optic neuropathy is useful for the diagnosis and early neurosurgical intervention. Delaying the diagnosis can lead to permanent visual loss.

Keywords:

Anterior clinoid mucocele, case report, compressive optic neuropathy

INTRODUCTION

Optic nerve lesions usually produce monocular visual loss, and in compressive optic neuropathies, gradual vision loss is the usual presentation. A variety of tumors (sellar and parasellar masses), dysthyroid ophthalmopathy, carotid-ophthalmic artery aneurysms, and abscesses can produce optic nerve compression. Mucocele in the anterior clinoid process is a rare cause of compressive optic neuropathy that can lead to monocular visual loss. Here, we describe a young female who presented with painless progressive monocular visual loss without any ophthalmoplegia, and magnetic resonance imaging (MRI) revealed an anterior clinoid process mucocele compressing the optic nerve.

CASE REPORT

A 19-year-old Sri Lankan girl presented with progressive, painless fluctuating right monocular

visual loss over a period of 1 year. Even though it was painless because of the fluctuating nature, the patient was initially treated with intravenous methyl prednisolone 1 g per day for 5 days at the local hospital before referral to a tertiary care center. There was no recordable improvement of vision following the steroid treatment. On admission to our ward, her right side visual acuity was 6/60 and left side visual acuity was 6/6. The right visual field was difficult to assess via the confrontation method and left visual field was normal. Fundoscopy revealed a pale disc on the right [Figure 1]. She did not have ophthalmoplegia but had a relative afferent pupillary defect on the right side.

Investigations including full blood count, liver function test, renal function tests, inflammatory markers, and thyroid function tests were normal. MRI revealed expansion of the right anterior clinoid process causing compression and kinking of the right optic nerve. The expanded anterior clinoid process showed internal signal abnormality (with both T1 and

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T2 showing intermediate high signal intensity), suggesting a mucocele formation. The connection of the clinoid process sinus with the right sphenoid sinus was obliterated facilitating mucocele formation [Figure 2]. There was no contrast enhancement [Figure 3]. Ocular tomography showed right-sided temporal retinal thinning [Figure 4]. Because the right side optic disc was already pale and retina was thinned, after discussing with the family members, the ophthalmology team, and the neurosurgery team, it was decided to manage conservatively. MRI was repeated 3 months later, and the mucocele did not show any interval change.

DISCUSSION

The Onodi cell is a posterior ethmoid cell which can pneumatize up to the sphenoid sinus and is intimately associated with the optic nerve and a mucocele of an Onodi cell causing optic neuropathy is extremely rare.^[1] These

mucoceles are nonneoplastic lesions and consist of mucus lined by a pseudostratified respiratory epithelium with a chronic inflammatory infiltrate.^[2] Mucocele can occur following mechanical obstruction of the pneumatizing air cell tract and the sinus ostium due to mucosal thickening, fibrosis, or bony overgrowth or following aberrant inclusion of ectopic mucinous tissue within the developing bone.^[3] The expansion and chronic inflammation can lead to gradual resorption and remodeling of the surrounding bone. Further enlargement of the mucocele may cause compression of adjacent structures.

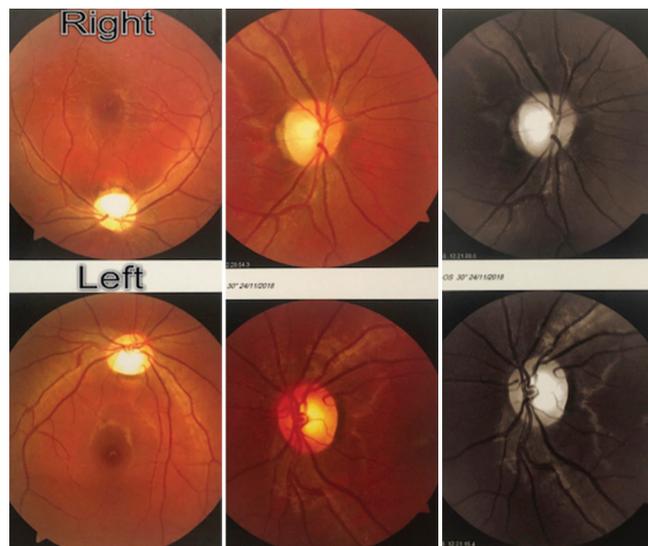


Figure 1: Fundal photography showing pale disk on the right side

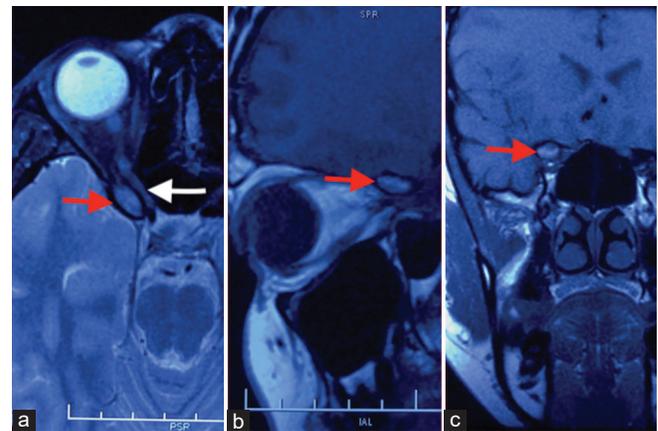


Figure 2: Magnetic resonance images of the right anterior clinoid process mucocele (red arrow) causing compression and kinking of the right optic nerve (White arrow). (a) T2-weighted/axial images. (b) Fluid-attenuated inversion recovery/sagittal images. (c) Fluid-attenuated inversion recovery/coronal images

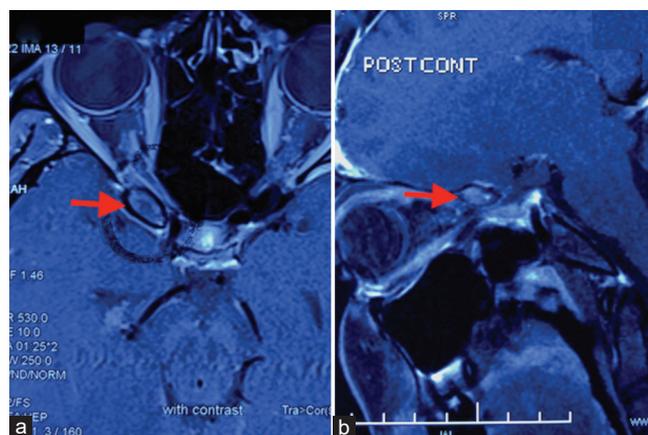


Figure 3: Postcontrast magnetic resonance images of the right anterior clinoid process mucocele (red arrow) showing no contrast enhancement. (a) T1 contrast = Axial images. (b) T1 contrast = Sagittal images

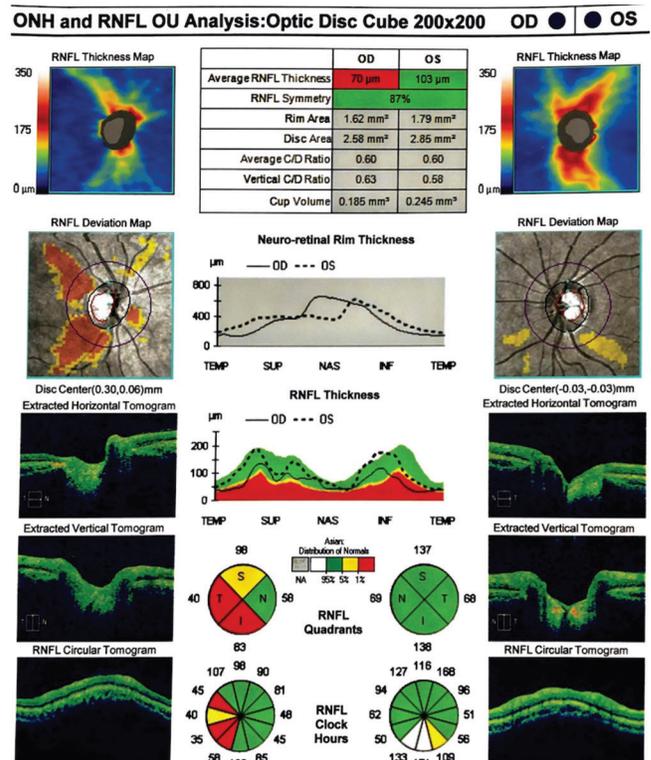


Figure 4: Ocular tomography showing right side temporal retinal thinning

Currently, there are only few case reports on mucocoeles involving the anterior clinoid process, leading to vision loss.

Some patients develop progressive visual impairment,^[4,5] and in some case reports, the vision loss was acute.^[6-10] Righini *et al.* reported a case of fluctuating monocular blindness as in our patient.^[11] This fluctuating nature can mimic inflammatory optic neuropathy, which can mislead the management. Our patient also received steroid treatment initially before the imaging studies suspecting inflammatory optic neuropathy. The fluctuating nature can be due to dynamic compression on the optic nerve, which varies with gradual resorption and remodeling of the surrounding bone. A rapid decline in visual acuity is generally considered a negative prognostic factor.^[2] Retrobulbar pain and painful visual loss were also the features in some patients in the literature.^[4,6,8,12,13] Dunya *et al.* reported a case of transient vertical double vision with vision loss due to an anterior clinoid process mucocoele.^[14] Forer *et al.* described a Chinese man of 50 years as having painful red eyes, ptosis, conjunctival injection, and complex ophthalmoplegia. In this case, MRI revealed a mucocoele between the optic nerve and carotid artery arising from anterior clinoid process.^[15]

The primary treatment for mucocoeles is surgical excision. Several approaches have been described for the surgery to remove the mucocoele including pterional craniotomy,^[4,8,16] endoscopic transsphenoidal,^[14] endoscopic transnasal approach,^[7,12] supraorbital craniotomy,^[5] transnasal sphenoid sinusotomy, endoscopic endonasal approach,^[10] and image-guided endoscopic transnasal approach.^[15] In some cases, steroids were also given as treatment.^[8,12,14] In most of the cases, complete recovery was seen with intervention.^[6,9,12,14,16] In our case, there was a delay of 1 year in diagnosing the patient, and on presentation, visual acuity was poor. Since the optic disc was already atrophic after a multidisciplinary team discussion, it was decided to manage conservatively without any neurosurgical intervention.

In conclusion, anterior clinoid process mucocoele is a rare cause of compressive optic neuropathy, leading to monocular visual loss. In this case, we need to highlight that the fluctuating nature of the visual impairment can mimic inflammatory optic neuropathy, which can mislead the clinicians. Some case reports also describe acute vision loss, retrobulbar pain, and painful visual loss as presenting complaints, which can further mislead. Early imaging is useful for the diagnosis in these cases, and early neurosurgical intervention can lead to complete recovery. However, delaying the MRI in resource-poor setting can lead to a delay in diagnosis and permanent visual loss, as in our case.

Consent to publish

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given consent for images and other clinical information to be reported in the journal. The guardian understands that the names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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